

## Neuromuscular Scoliosis: A Neurological Point of View

Celmir de Oliveira Vilaça<sup>1,2</sup>, Jacqueline Stephanie Fernandes do Nascimento<sup>3</sup>, Nicolle dos Santos Moraes Nunes<sup>3</sup>, Thais de R. Bessa-Guerra<sup>3</sup>, Marco Antonio Alves Azizi<sup>4</sup>, Adalgiza Mafra Moreno<sup>5</sup>, Antônio Marcos da Silva Catharino<sup>6</sup>, Paulo Henrique de Moura<sup>4</sup>, Thiago Rodrigues Gonçalves<sup>4</sup>, Renata R. T. Castro<sup>4</sup>, Marcos RG de Freitas<sup>4</sup>, Marco Orsini<sup>4</sup>.

<sup>1</sup>Institute of Traumatology and Orthopedics (INTO), Rio de Janeiro-RJ, Brazil.

<sup>2</sup>University Hospital Clementino Fraga Filho (HUCFF-UFRJ), Rio de Janeiro-RJ, Brazil

<sup>3</sup>Graduanda de Medicina na Universidade Iguazu (UNIG) – Nova Iguaçu, RJ –Brasil.

<sup>4</sup>Professor titular na Universidade Iguazu (UNIG). Nova Iguaçu, RJ –Brasil.

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**Abstract** — *Objective: make a review of the main neurological diseases associated with neuromuscular scoliosis and perioperative care during scoliosis correction surgeries. Method: Non-systematic review using a Google Scholar platform for articles with the descriptor “neuromuscular scoliosis” in the journal title. Results and Discussion: 46 articles in English and 3 articles in Portuguese were used for this review. Conclusion: Neuromuscular scoliosis has the fastest course of evolution, higher rates of disability and higher rates of complications in corrective surgery when compared to idiopathic scoliosis. The knowledge of neurological diseases more associated with neuromuscular scoliosis helps in its early detection and its long-term monitoring.*

### I. INTRODUCTION

We can define scoliosis as a three-dimensional deformity of the spine, including a curvature in the frontal plane greater than 10° (ten degrees). This curvature is assessed by measuring the Cobb's angle.<sup>1,2</sup>

Cobb's angle was first described by John R. Cobb. It is obtained on panoramic radiographs of the spine and calculated by the angle of intersection between the upper plateau of the upper vertebra and the lower plateau of the lower vertebra of the scoliotic curve (figure 1).<sup>3</sup> Scoliosis can be divided into idiopathic (IS), congenital and neuromuscular scoliosis (NMS). Idiopathic scoliosis is subdivided into infantile, juvenile and adolescent forms. IS develops without the presence of a detectable disease as a

cause of progressive spinal curvature.<sup>2</sup> Several theories try to explain idiopathic scoliosis through genetic factors or even hormonal imbalance, such as changes in melatonin levels.<sup>4</sup> IS does not usually maintain its progression during adulthood, but it may need surgical correction in order to avoid the appearance of cardiopulmonary dysfunctions or pelvic obliquity with consequent difficulty in maintaining posture.<sup>5</sup> Congenital scoliosis, on the other hand, is the most frequent form of congenital spinal deformity, overcoming congenital kyphosis or lordosis. It is due to some aggression to the fetus during the embryological development of the spine and is often associated with other malformations, be they cardiac, urinary or gastrointestinal tract.<sup>6</sup>

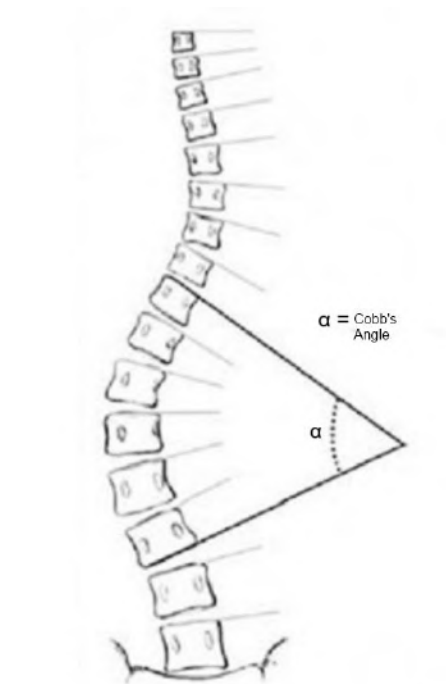


Fig.1: Cobb's angle for scoliosis evaluation.

On the other hand, neuromuscular scoliosis is a scoliotic deformity secondary to muscle imbalance caused by neuropathic or myopathic diseases.<sup>7</sup> NMS generally has an earlier onset and faster progression when compared to idiopathic scoliosis. It also has higher hospitalization costs, operative complications and number of surgeries for correction.<sup>8,9</sup> Another difference between idiopathic and neuromuscular scoliosis is the pattern of the scoliosis curve. In IS there is a convex scoliosis curve generally to the right, sometimes with the presence of a double curve whereas in NMS scoliosis is type C with a large single curvature affecting the thoracic and lumbar spine.<sup>10,11</sup>

The article aims to recognize and discuss the neurological disorders most often associated with NMS. The neurologist is sometimes the first to assess these patients and one of those responsible for their long-term care. Early diagnosis by the neurologist can positively impact the prognosis of individuals with NMS.

## II. METHODS AND RESULTS

We performed a non-systematic review of articles through the Google Scholar platform from January 1992 to October 2020. We prefer articles in Portuguese or in English whose title had the descriptor "neuromuscular scoliosis" or "escoliose neuromuscular" in the journal title. We obtained a total of 524 articles from this search. We include experimental, review articles, case reports or expert opinions. Some papers were chosen from the references obtained from the articles of the initial search. Others were

chosen because they address the neurological diseases most often responsible for NMS. We used a total of 49 articles for this manuscript, 46 in English and 3 in Portuguese language.

## III. DISCUSSION

### Definition and Epidemiology of Neuromuscular Scoliosis

Neuromuscular scoliosis can be defined as a non-congenital spine deformity occurring in individuals with a diagnosis of pre-existing neuromuscular disease.<sup>12</sup> While in neurology the term neuromuscular generally describes only pathologies associated with diseases of the peripheral nervous system with impairment of the muscle or motor neuron, orthopedists use the term for any neurological disease, including cerebral palsy.<sup>5</sup> We can detect the presence of scoliosis in individuals with neurological disease by performing the Adams test or the anterior tilt test.<sup>13</sup> The individual bends forward with arms forward, palms facing each other and with feet together. The presence of asymmetries in the tangential visualization of the paravertebral musculature indicates a positive test and the need for radiographic testing to analyze the Cobb's angle.<sup>13,14</sup>

NMS can occur in up to 90% of children with neuromuscular disease and progress after adulthood regardless of the maturity of the skeletal system, being generally refractory to conservative treatments.<sup>15</sup> The first cases of NMS with surgical correction were described in individuals with poliomyelitis.<sup>16</sup> Currently among the neurological conditions most often triggering NMS are cerebral palsy, Duchenne Muscular Dystrophy, Spinal Muscular Atrophy and Friedreich's ataxia.<sup>12</sup>

### Neurological Diseases Most Associated with Neuromuscular Scoliosis

#### A) Cerebral palsy

Cerebral palsy (CP) can be defined as the presence of motor dysfunction during birth due to an identifiable brain injury during pregnancy. It affects seven out of every 1000 live births in developing countries.<sup>17</sup> It can be subdivided into spastic, choreoatetoid, non-toxic or mixed types. CP is one of the main causes of neuromuscular scoliosis, especially in its quadriplegic spastic form, followed by diplegic and hemiplegic forms.<sup>18,19</sup> The incidence of NMS in individuals with CP depends on the definition used for scoliosis, but rates between 25 and 69%.<sup>19,20</sup> Diseases with hypertonia of the trunk muscles such as CP are more often associated with the presence of NMS when compared to muscle diseases that cause hypotonia of the paraspinal muscles.<sup>21</sup> The risk of developing neuromuscular scoliosis

in individuals with CP increases with age or degree of motor disability due to the classification of gross motor function, being higher in grades IV and V (table 1).<sup>22-24</sup>

Table 1 - Gross Motor Function Classification System

LEVEL 1	Walks without restrictions, with limitations for more complex motor activities (running, jumping)
LEVEL 2	Walks without assistance, but with limitations in community gait
LEVEL 3	Walks with support, with limitations in the community walk
LEVEL 4	Mobility is limited, requires a wheelchair in the community
LEVEL 5	Severely limited mobility even with the use of assistive technology

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### B) Duchenne Muscular Dystrophy

Duchenne muscular dystrophy (DMD) affects two to three boys in 10,000. It is a recessive disease characterized by the presence of a mutation in the dystrophin gene at position 21 of the short arm of the X chromosome. Dystrophin is a protein responsible for maintaining calcium homeostasis.<sup>25</sup> Individuals with Duchenne often have progressive scoliosis in the first decade of life. NMS is found in up to 90% of patients with DMD.<sup>26</sup> The lordotic posture in patients with Duchenne and the ability to walk can delay the development of scoliosis and corticosteroid therapy can decrease scoliosis rates with the need for surgical correction from 90 to 15% of cases.<sup>16</sup> Deflazacort, an oxazolone derivative of prednisone, had slowed the progression of scoliosis with lower rates of long-term use compared to other corticosteroids.<sup>27</sup> There are defending authors of early corrective scoliosis surgery in mild cases to preserve respiratory function, while others perform surgery only in severe cases because not all cases of DMD evolve to severe scoliosis. Furthermore, surgery sometimes does not increase survival despite the positive impact on daily activities.<sup>26</sup>

### C) Spinal Muscular Atrophy

Spinal muscular atrophy (SMA) represents a recessive autonomic genetic disease with muscle atrophy due to degeneration of the motor neurons in the anterior spinal cord and presents in eight out of every 100 thousand live births.<sup>28</sup> This degeneration occurs due to deficiency of the neuronal survival protein due to mutations in the SMN 1 gene. There are three types of SMA in childhood (I-III) based on the age of onset and motor development milestones.<sup>29</sup> NMS occurs mainly in type II spinal muscular atrophy, with rapid progression associated with pulmonary restriction.<sup>1</sup> Scoliosis rates in AMS are around 80% of cases.<sup>10</sup> Unlike individuals with DMD where ventilatory restriction affects the diaphragm, individuals

with type II SMA have pulmonary restriction due to predominant deformity in the thoracic region, causing the parasol/ umbrella chest phenotype.<sup>30</sup> In SMA Type III only occurs NMS later, after loss of ambulation.<sup>31</sup>

### D) Friedreich's ataxia

It is the most common form of progressive spinocerebellar degeneration. It corresponds to a recessive genetically transmitted ataxia, caused by repetition of GAA trinucleotides (guanine, adenine, adenine) within the frataxin gene on chromosome 9q13.<sup>32-34</sup> The mutation in the frataxin gene causes iron accumulation in the mitochondria, with cell death.<sup>32</sup> Clinically, it presents as a progressive cerebellar ataxia beginning at around 25 years of age, associated with peripheral neuropathy and pyramidal symptoms. Other changes can also be found, such as pes cavus, optic atrophy, diabetes and cardiomyopathy.<sup>33</sup> NMS occur in about 60% of patients with Friedreich and in 30% of cases scoliosis is not progressive. The younger the age of onset of symptoms, the more frequent is the presence of scoliosis and this does not depend on gait changes.<sup>33</sup>

### E) Myelomeningocele

It is the main type of spinal dysraphism and can cause NMS both by injury to the first motor neuron and by injury to the second neuron at the anterior tip of the spinal cord. The cord and nerves protrude through the closed LAMINA in an incomplete way.<sup>1</sup> The prevalence of scoliosis in patients with myelomeningocele is 53%, with a slight predominance in females.<sup>35</sup> Complication rates in myelomeningocele surgeries may be more frequent when compared to other causes of neuromuscular scoliosis, highlighting high rates of urinary tract infection.<sup>8</sup>

### F) Charcot-Marie-Tooth disease

Also known as hereditary sensory-motor neuropathy, Charcot Marie-Tooth Disease (CMT) is the main hereditary peripheral neuropathy. It can be divided into dominant, recessive and X-linked forms.<sup>36</sup> Although it most commonly affects the limbs, especially the lower ones, it is capable of causing NMS in up to a third of cases. CMT is best known for being the leading cause of pes cavus of neurological origin in the general population.<sup>36,37</sup>

### G) Other conditions

Poliomyelitis, although it is the first disease where the issue of scoliosis in neuromuscular diseases has been raised, is currently a condition rarely found due to vaccination.<sup>38</sup> Guillain-Barré syndrome has rare reports of scoliosis as a sequel.<sup>39</sup> Congenital myopathies, although rare and non-progressive, can also cause scoliosis over the years.<sup>40</sup>

### Preoperative Evaluation in Neuromuscular Scoliosis

The main indication for scoliosis surgery is the presence of curvature of the trunk or pelvic obliquity with inability to maintain posture and sitting position.<sup>12</sup> Cobb's angle greater than 40° may also indicate scoliosis with necessity of surgical correction.<sup>18</sup> Pelvic obliquity is defined as the failure of the pelvis to remain perfectly horizontal in the frontal plane. The risks of pelvic obliquity are greater in patients with NMS of spastic origin such as CP when compared to the flaccid forms found in DMD and AMS.<sup>12</sup> Despite the risks of extensive surgery in debilitated individuals, patients undergoing scoliosis correction surgery have improved quality of life, lower pain rates and higher levels of family satisfaction after the procedure.<sup>18,24,41</sup>

Unlike cases of IS, individuals with NMS may present cardiac involvement due to their neurological condition.<sup>14,32</sup> Patients with DMD have heart disease in 10 to 20% of cases. Undiagnosed cardiomyopathy in DMD associated with blood loss during scoliosis correction surgery can lead to perioperative cardiac arrest. DMD cardiomyopathy sometimes does not correlate with echocardiographic dysfunction, and a cardiac study by magnetic resonance is suggested.<sup>14</sup> Individuals with Friedreich's Ataxia present with hypertrophic cardiomyopathy with left ventricular dysfunction and this can progress to dilated heart disease, contraindicating scoliosis surgery.<sup>32</sup>

Considering patients with CP, it is important in the perioperative period to evaluate the adjustment of medications against spasticity. Drugs such as baclofen can induce seizures during both their introduction and withdrawal.<sup>42</sup> In addition, there is controversy as to whether individuals with CP can present scoliosis progression with the use of baclofen through an intrathecal pump.<sup>43</sup>

### Postoperative Complications in Neuromuscular Scoliosis

Complication rates in neuromuscular scoliosis surgeries are estimated to range from 18 to 75% in the postoperative period. These complications are classified in different ways. One of these classifications divides complications into intraoperative and early and late postoperative.<sup>11</sup>

Respiratory disorders are the most frequent postoperative complications in most studies and most observed in individuals with forced vital capacity below 50% in the preoperative period. In these cases, noninvasive ventilation before surgery can decrease postoperative risks, especially when vital capacity is below 40% of predicted values.<sup>44</sup>

Gastrointestinal disorders are attributed to paralytic ileus

by medications such as opioids or during anesthetic procedures, presence of gastroesophageal reflux or in patients with poor nutritional conditions, with high risk of superior mesenteric artery compression syndrome.<sup>45</sup> In debilitated patients below the fifth weight percentile, nutritional support is recommended weeks to months before surgery.<sup>9</sup>

Infections after the surgical procedure are associated with high or low body mass index, lymphocyte count below 1500 mm<sup>3</sup> or serum albumin levels below 3.5 g/dL.<sup>45</sup> Among the agents most involved in the infection of surgical wounds are *S. Aureus*, *P. aeruginosa* and *E.coli*.<sup>46</sup> It is important not to confuse fever of infectious origin with fever due to the release of inflammatory cytokines such as IL-6 and TNF- $\alpha$  from the extensive surgical procedure to correct scoliosis. This febrile condition occurs on average up to seven days after the procedure in up to 20% of cases.<sup>15</sup>

NMS surgeries show greater volumes of bleeding when compared to individuals with IS and different etiologies of NMS may show different volumes of blood loss. For example, individuals with DMD have higher rates of bleeding compared to individuals with AMS.<sup>47</sup> This is explained to the absence of dystrophin in the smooth muscle cells of the vessels and the consequent contractile inability to stop the bleeding.<sup>25</sup> Individuals with CP and associated epilepsy, on the other hand, may have a greater volume of bleeding due to platelet dysfunction or coagulation factors deficiency from the use of anticonvulsants.<sup>9</sup> Hypotension during surgery can reduce blood loss, however it can increase the risk of neurological injury. An alternative to a drop in hematocrit may be to transfuse autologous blood taken before the procedure and avoid increased abdominal pressure over the vena cava for long periods in the decubitus position.<sup>48</sup> It is possible to reduce the risk of complications by performing scoliosis correction surgery in more than one procedure in different days, however at the expense of longer hospitalization. Finally, tranexamic acid, a substance that inhibits fibrinolysis, can be used during scoliosis surgery to decrease bleeding rates.<sup>47</sup>

### Intraoperative Monitoring in Neuromuscular Scoliosis Surgery

Neurophysiological monitoring is useful during surgeries to correct idiopathic scoliosis and, despite its high cost, can be compensated by a reduction in neurological sequelae and length of hospital stay. It mainly uses the somatosensory evoked potential (PESS) by measuring the fall in its amplitude or velocity of conduction.<sup>49</sup> In NMS can be difficult to assess the PESS as dysfunction often affects the underlying nerve conduction as in Friedreich's



ataxia.<sup>32</sup> However, some pathologies associated with NMS may present a low risk of injury during scoliosis surgeries regardless of the use of intraoperative monitoring as in cases of CMT.<sup>37</sup> Another possibility of operative monitoring in patients with NMS is through motor evoked potential by transcranial electrical stimulation. However, there are risks of inducing seizures in susceptible individuals, such as patients with CP.<sup>49</sup> The combined use of PESS and transcranial electrical stimulation can increase the reliability of intraoperative neurophysiological monitoring.<sup>44</sup> The Stagnara test or awakening test can be difficult to perform in individuals with neuromuscular scoliosis and intellectual dysfunction in the absence of intraoperative monitoring.<sup>45</sup> The test consists of reducing sedation and mobilizing the lower limbs during surgery. Failure to mobilize may indicate neurological injury requiring suspension of the procedure.<sup>49</sup> Finally, although more frequent compared to IS surgery, the incidence of neurological injuries during NMS surgery is lower than other operative complications.<sup>8</sup>

#### IV. CONCLUSION

Neuromuscular scoliosis is a clinical condition resulting from several common pathologies in neurological clinical practice. These illnesses can cause scoliosis through different pathophysiological mechanisms. NMS presents faster progression, frequent requirement of surgical correction and high rates of complications in the perioperative period when compared to idiopathic scoliosis. Under the perspective of multidisciplinary health care, scoliosis surgery is performed by the orthopedic surgeon. However, the neurologist usually participates in the assessment regarding the etiological diagnosis of NMS or in the follow-up of these individuals regardless of the indication for surgical correction.

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